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CASE REPORT

Fibrous dysplasia with secondary aneurysmal bone cyst

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Case report

A 33-year-old woman with known polyostotic fibrous dysplasia involving the right upper extremity presented with right elbow pain and swelling of rapid onset. Her past medical history was remarkable for a pathologic fracture of the right humerus treated by intramedullary rod fixation. There was no evidence of McCune-Albright's syndrome. Laboratory tests were normal.

Radiographs of the right elbow demonstrated a rapid 3-month-interval change with lytic expansion of the proximal radius (Fig. 1). Skeletal scintigrams revealed a defect of radionuclide uptake at the previously ^{99}Tc-MDP-avid site of radial fibrous dysplasia. Magnetic resonance (MR) imaging showed a "blow out" proximal radial lesion with a signal isointense to the forearm muscles on T1-weighted images (Fig. 2A, B). A T2-weighted spin echo sequence demonstrated a lobulated mass containing multiple fluid-fluid levels within a surrounding dark rind (Fig. 2C). Concern about malignancy prompted an open biopsy.

A frozen section of the biopsy specimen demonstrated no evidence

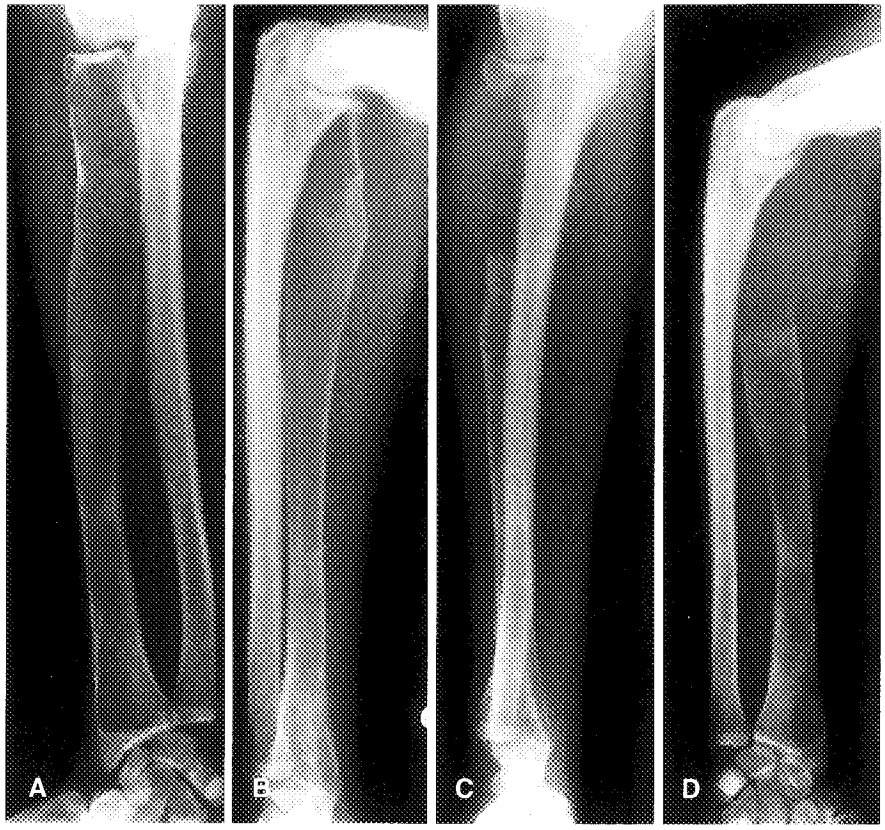

Fig. 1A–D Two sets of radiographs of the right forearm obtained 3 months apart. **A, B** The initial radiographs showed cortical thinning and minimal expansion of the proximal radius with a ground glass pattern characteristic of fibrous dysplasia. **C, D** Follow-up radiographs 3 months later, after onset of forearm tenderness and swelling, demonstrated a marked expansile and lytic lesion of the proximal radius. The cortical bone was markedly thinned secondary to the expansile process

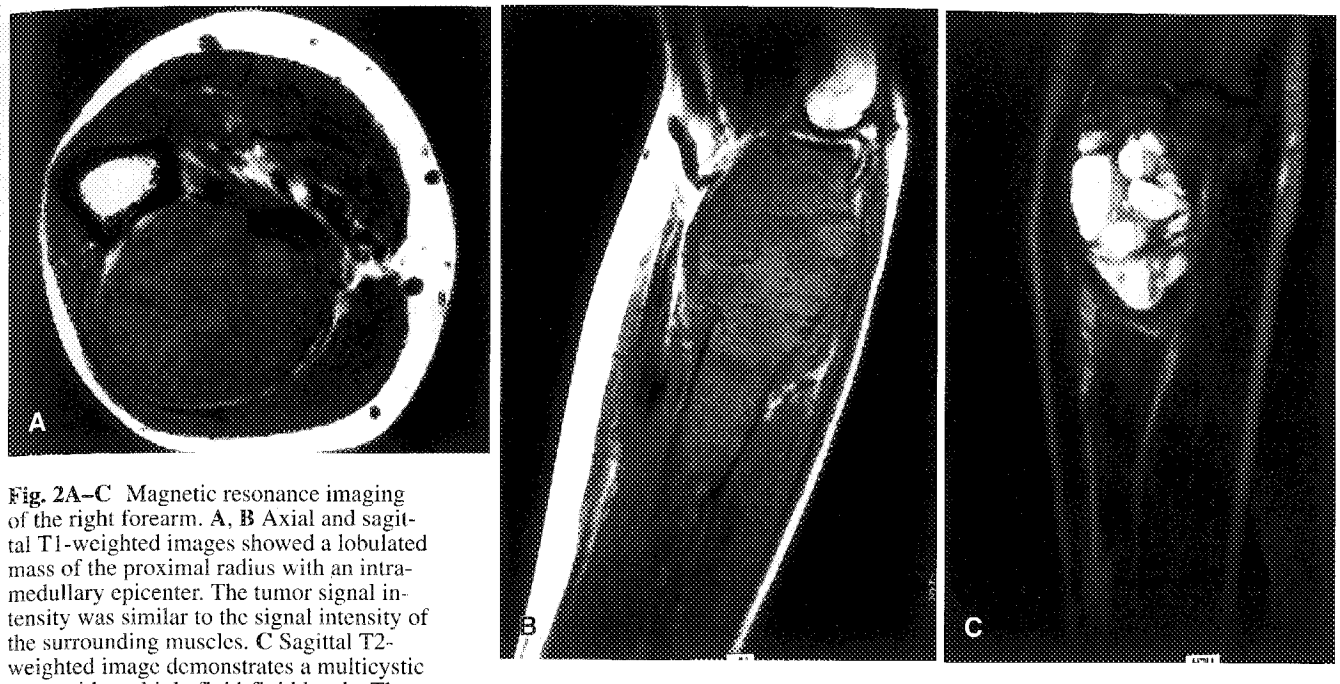

Fig. 2A–C Magnetic resonance imaging of the right forearm. **A**, **B** Axial and sagittal T1-weighted images showed a lobulated mass of the proximal radius with an intramedullary epicenter. The tumor signal intensity was similar to the signal intensity of the surrounding muscles. **C** Sagittal T2-weighted image demonstrates a multicystic mass with multiple fluid-fluid levels. The tumor was delineated by a lobulated dark rind

of malignancy. The lesion was curetted and the remaining periosteal shell was collapsed upon itself. The specimen showed typical features of fibrous dysplasia with a spindle cell fibrous tissue background and woven bone in a "Chinese letter" pattern (Fig. 3). Cystic blood-filled spaces with fibrous tissue lining were noted, representing focal aneurysmal bone cyst transformation (Fig. 4). Fibrous dysplasia with secondary aneurysmal bone cyst was diagnosed. The differential diagnosis on the basis of the radiographic findings included malignant sarcomatous transformation of fibrous dysplasia, pathologic fracture with hematoma, and aneurysmal bone cyst change in fibrous dysplasia.

The patient's postoperative course was unremarkable.

Discussion

Fibrous dysplasia is a developmental disorder characterized by fibrous osseous metaplasia of the medullary cavity [1–3]. Its manifestation can be monostotic or polyostotic. The polyostotic form is six times less frequent, can be monomelic, and has a slight predilection for female patients in the first and second decades of life [2, 3]. Any bone can be involved in polyostotic fibrous dysplasia. The most common locations are the lower extremity and pelvis [4]. Occurrence at the upper extremity is moderately frequent. Polyostotic fibrous dysplasia may be associated with McCune-Albright's syndrome with cutaneous pigmentation, endocrine disorders, precocious puberty, and bone deformity. It has a more aggressive clinical course than the monostotic form and pathologic fractures are common. Malignant transformation occurs in less than 1% of all cases of fibrous dysplasia [5]. Pain, swelling, and significant interval change of the involved bones are warning signs of malignancy [2]. Osteogenic sarcoma, fibrosarcoma, chondrosarcoma, and giant cell sarcoma, in decreasing order of frequency, are the principal forms of neoplastic degeneration. The sarcomatous lesions have a poor prognosis, with a 5-year survival rate of 52% after radical surgery [4].

Histologically, fibrous dysplasia consists of bland fibrous tissue stroma with spicules of woven bone from fibrous osseous metaplasia. The bone spicules may demonstrate a curled "C" or "Y" pattern resembling Chinese characters or "alphabet soup" [1, 4]. Gross pathology demonstrates a firm, whitish, and gritty tissue with loss of normal bone trabeculae [6]. The lesion originates from the intramedullary region, frequently eroding and expanding the cortex. Cystic degeneration could be encountered with multiple loculated clear fluid-filled spaces containing a monolayer cell lining.

Plain radiograph and computed tomographic features of fibrous dysplasia reflect the woven bone production that is characteristic of this disorder [1, 2]. The lesion, with an intramedullary epicenter, has a characteristic ground glass pattern and demonstrates different degrees of X-ray attenuation related to the amount of mineralization. The fibrous dysplastic focus is usually well-marginated and surrounded by a sclerotic rind [1]. The lesion may be expanded secondary to cystic degeneration or aneurysmal bone cyst for-

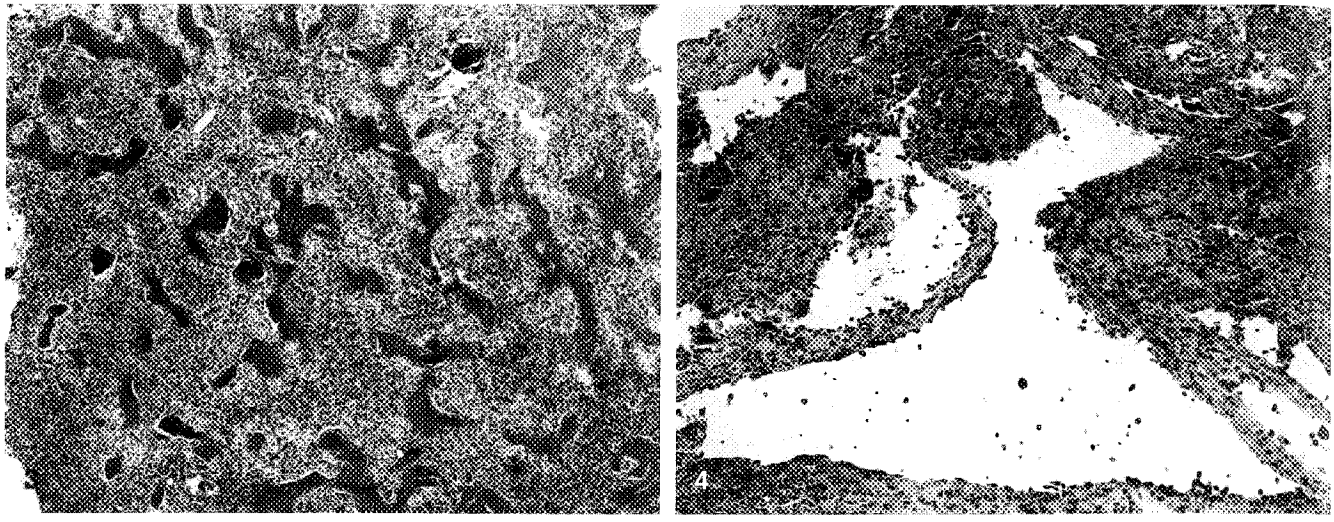

Fig. 3 Photomicrograph showing an area of typical fibrous dysplasia. There is a spindle cell fibrous tissue background with woven bone in a "Chinese letter" pattern. (H&E, ×82)

Fig. 4 Photomicrograph showing the fibrous tissue lining of a cystic space, consistent with aneurysmal bone cyst transformation. (H&E, ×102)

mation. Cortical destruction and soft tissue involvement may be encountered in cases of pathologic fracture and in rare instances of sarcomatous transformation. Bone scintigraphy demonstrates increased radionuclide uptake in involved sites [2, 3]. MR imaging shows a lesion with signal isointense to the surrounding muscular layers on T1-weighted sequences and a variable signal intensity on T2, depending on the metabolic status of the fibrous dysplastic tissue [3]. Fluid-fluid levels have been demonstrated on MR as characteristic of cystic degeneration in fibrous dysplasia [5, 7]. This feature, representing serous and/or bloody fluid layering within septated cavities, is, however, nonspecific.

The differential diagnosis includes a large spectrum of malignant and benign conditions such as telangiectatic osteosarcoma, malignant fibrous histiocytoma, simple bone cyst, and aneurysmal bone cyst [7]. This last entity is a known complication of fibrous dysplasia and may show rapid expansion that mimics a malignant tumor. This is specially true in the skull and face [6, 8, 9]. Aneurysmal bone cyst is composed of multicystic blood-filled fibrous tissue with three developmental stages: a permeative or circumscribed lytic early phase, a malignant-appearing "blow out" midphase, and a late equilibrium phase with a well-marginated periosteal rim [6]. Aneurysmal bone cyst may occur at any time during life but it is more common during the second and third decades [9]. It can occur anywhere in the skeletal system; however, the long bones of the extremities are more often involved than are the skull and facial bones [8, 9]. Approximately 30% of aneurysmal bone cysts are considered a secondary process in the presence of other primary lesions such as fibrous dysplasia, chondroblastoma, chondromyxoid fibroma, osteosarcoma, and giant cell tumor. The remaining 70% occur as primary lesions [10]. However, aneurysmal bone cysts may not be considered true neoplasms [8, 9], since they may arise from anomalous hemodynamic conditions of preexisting lesions, and sometimes obliterate the primary tumor with their exuberant growth.

The case presented here is an example of known fibrous dysplasia in the appendicular skeleton that over a short time showed a radiographic pattern of rapid growth. The preoperative diagnosis was possible malignant transformation. However, the pathologic diagnosis was focal aneurysmal bone cyst transformation. This case suggests that aneurysmal bone cyst engraftment on fibrous dysplasia may masquerade as malignant transformation. If this association is considered, radical surgery may be avoided. The MR features are nonspecific and based on fluid-fluid levels. Sarcomatous transformation of polyostotic fibrous dysplasia, secondary midphase aneurysmal bone cyst, simple cystic degeneration of fibrous dysplasia, or a combination of these three processes could fit the MR findings. The dark rind surrounding the bulky proximal radial lesion may be the only clue of benignity [5]. Polyostotic fibrous dysplasia is a benign developmental disorder that may be associated with bone deformity, pathologic fracture, secondary aneurysmal bone cyst, cystic degeneration, and sarcomatous transformation. Any of these complications can demonstrate bone expansion and cortical thinning or disruption with soft tissue involvement. Plain radiographs and MR, in this case, were helpful in assessing the evolution of the lesion. However, the MR findings are nonspecific regarding tumor behavior. Aggessive benign processes (aneurysmal bone cyst formation) cannot always be distinguished from malignant change.

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